431* A novel method of projecting cystic fibrosis birth cohort survival estimates that overcomes short duration of follow-up

A. Jackson[1,2], L. Daly[1], C. Kelleher[1], A.L. Jackson[3], M. Harrington[2], S. Zhou[2], L. Foley[2], P. Fitzpatrick[1]. [1]*University College Dublin, School of Public Health, Physiotherapy & Population Science, Dublin, Ireland;* [2]*Cystic Fibrosis Registry of Ireland (CFRI), Woodview House, Ireland;* [3]*Trinity College Dublin, School of Natural Science, Dept of Zoology, Dublin, Ireland*

The current lifetable method, which is frequently used to estimate CF life expectancy, does not allow for changes in age-specific mortality rates over time despite observed improvements in childhood mortality rates. Longitudinal survival analysis of birth cohorts facilitates rate adjustment, but median survival estimates may not be available for many years.

We aimed to improve CF survival analysis by developing a parametric modelling technique to estimate birth cohort survival in situations where duration of follow-up is short.

Data on births and registered deaths were sourced from the CFRI (>90% enrolment by 2009) and the Central Statistics Office, 1980–2007. Current lifetable estimates were prepared using the US registry method and the Kaplan–Meier (K–M) method was employed to calculate survival in 4 five-year birth cohorts (1980–1999). The best fitting parametric model to birth cohort data was selected.

Current lifetable estimates of annual median survival fluctuated from year to year, requiring a lengthy observation period to establish a trend of increased survival. K–M estimates showed increased survival in successive birth cohorts, due to greater proportions surviving the first year of life. Gender and year of birth had an independent effect on survival. Median survival was unknown for persons with CF born 1985–94 and surviving the first year of life because 85% were alive in 2007. Using a Weibull regression model, we estimated a median age at death of 39.4 years (95% CI: 31, 47.8) for this cohort.

This novel method deals effectively with survival estimation where the duration of birth cohort follow-up is short, and the age profile of decedents continues to evolve.

432 Survival analysis of cystic fibrosis (CF) patients in the North Al Bathina region of the Sultanate of Oman

U. Fass[1], M. Al Oraimi[1], R. Thomas[1], S. Dhouwayani[1], M. Al Naqbi[1], N. Al Balushi[1], F. Al Hinai[1], G. Shivalingam[2], F. Al Balushi[1], A. Rajeev[1], T. Heming[1]. [1]*Oman Medical College, Human Function/Biochemistry, SOHAR, Oman;* [2]*Sohar Regional/Teaching Hospital, SOHAR, Oman*

The survival and morality data of CF patients depend on the severity of the disorder, interventions by effective patient care, and patient compliance. CF patient registries do not exist in the Arab population, so information about the survival of Arab CF patients is largely unknown. We identified 39 CF patients (17 male, 21 female, age 1–15 yr) born between 1992 and 2009 by a complete retrospective and prospective data collection in the North Al Bathina region of the Sultanate of Oman. Since patients were alive at the end of the study period, we utilized a right censored product-limit estimation for the survival and mortality analysis. The variants of the survival probability were analyzed by Greenwood's variance expression. Normal like distribution was achieved by logarithmical transformation of the survival probability. The mean survival time estimate is 10.5 years with a normal-based approximate 95% confidence interval of ±1.8 years. The median survival time estimate of 10±1.5 years is similar to the mean survival time estimate. Interestingly, the mean survival time estimate of female CF patients in our cohort is significantly lower than of the male patients (8.7 years and 13.3 years retrospectively). This observation agrees with the know gender difference in CF mortality. The study documents a very low life expectancy of CF patients in our cohort. This may result from a combination of factors, e.g. genetic predisposition, lack of multidisciplinary management, or missed early diagnosis.

433* Geriatric cystic fibrosis: clinical features of patients over 50 years of age

D. Hadjiliadis[1], M. Kaplan[2], M. Ferrin[2], G. Imbesi[3], D. Dorgan[2], D. Holsclaw[4]. [1]*University of Pennsylvania, Medicine, Philadelphia, PA, United States;* [2]*University of Pennsylvania, Philadelphia, PA, United States;* [3]*University of Pennsylvania, Pulmonary, Allergy and Critical Care, Philadelphia, PA, United States;* [4]*University of Pennsylvania, Philadelphia, PA, PA, United States*

Introduction: Median survival of cystic fibrosis (CF) has been steadily increasing approaching 40 years. As a result, a significant number of patients are over 50 years old and have unique features in their diagnosis, presentation and clinical problems compared to patients with classic CF. We aim to describe this population in our clinic.

Methods: Patients over 50 years of age from 2002 to 2008 in our clinic were included. Diagnostic characteristics, genetic data, clinical features, microbiological features and other comorbidities were collected.

Results: There were 30 patients that met the inclusion criteria. Nineteen patients were female (63.3%) and twenty-eight were white (93.3%). Two patients (6.7%) received lung transplant before their 50th birthday. Average at diagnosis was 26.1±20.7 years and 16/30 (53.3%) were diagnosed in childhood or adolescence. Only two patients *(6.7%)* had meconium ileus. Mean sweat chloride was 100.1 mM. Genetic testing revealed five (16.6%) patients who were DF508 homozygotes, while the prevalence of DF508 was 40% (24/60 genes genotyped). Pseudomonas was present in 21 (70%) patients at last follow-up. At last follow-up, mean FEV1 was 1.45 L and FVC 2.29 L, while mean BMI was 23.1. Eight (26.7%) patients had passed away at the time of this report. Eleven (36.7%) patients had not received IV antibiotics during the study period. Diabetes was common in this group.

Conclusions: Patients with CF reaching 50 years of age have significant burden of disease even though many of them are diagnosed as adults. Future registry studies should describe further this group and its needs

434* "Old" patients with CF: report from Brittany (Western France) where CF is frequent

V. Scotet[1], I. Duguépéroux[1], M.-P. Audrézet[1,2], G. Rault[3], S. Ramel[3], R. Michel[4], V. Moisan-Petit[5], C. Férec[1,2]. [1]*Inserm, U613, Brest, France;* [2]*Hospital, Dep of Molecular Genetics, Brest, France;* [3]*CRCM, Roscoff, France;* [4]*CRCM, Rennes, France;* [5]*CRCM, Vannes, France*

As a result of the improvement in survival of people with CF, the proportion of "old" patients is increasing. This study aimed to describe the characteristics of CF patients born in Brittany who survived the age of 35.

Active enquiries were done to identify patients with classical form of CF, aged of 35 y. or more who are still alive, as well as those who died after the age of 35. Patients were recruited through the registry set up in Brittany and by contacting the CF centres and pulmonologists of the area. Vital statute was updated through CF centres and birth councils.

A total of 53 long-term survivors (28 males, 25 females) were registered; 47 (68.8%) of them were still alive at the ending date (Dec 31st 2008). Mean age of alive patients was 41.4 y. (the oldest one being 59.9 y. old) whereas that of dead patients was 37.8 y. The spectrum of *CFTR* mutations differed from that classically observed in CF; the 2789+5G>A being particularly frequent (8.5% vs. 0.6%, $p < 0.0001$).

As our registry enrolled alive and dead patients born in Brittany since 1970, we were also able to estimate, using Kaplan-Meier method, the survival probability of patients born between 1970 and 1974 ie. likely to have reached the age of 35. That cohort included 78 patients among whom 3/4 have died so far (n = 59). Median survival was estimated at 17.1 y. (95% CI: [10.3; 25.6]). It did not differ between gender ($p = 0.775$) and was lower than in the younger generations (log-rank: $p = 0.0086$).

This study confirmed the genotypic specificities of long-term CF survivors and was able to precisely assess median survival of patients born in the 1970s.

Acknowledgments: Practitioners who contributed to data, P.H.R.C.